Portal Hypertension Secondary to Choledochal Cyst

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Two patients with portal hypertension secondary to choledochal cyst are reported and combined with four others previously reported in the literature. Choledochal cysts may, by extrensic compression, cause partial or complete portal vein obstruction, portal hypertension and esophageal varices. Needle biopsy of the liver is probably not a reliable means of differentiating choledochal cyst from other intrahepatic causes of portal hypertension. Internal drainage of the choledochal cyst has been performed in four patients and in each instance resulted in satisfactory portal decompression.

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PORTAL HYPERTENSION as a complication of choledochal cyst has been documented in four of more than 650 previously reported cases of choledochal cyst.[1,5-8] The etiology of the portal hypertension in each instance was not readily apparent and often led to delay in arriving at the complete diagnosis. During the past ten years, two of the eight children admitted to our hospital with choledochal cyst were found to have associated portal hypertension. Our experience combined with that previously reported suggests significant conclusions regarding both diagnosis and management.

Case Material

Case 1. An eight month old girl was admitted to Children's Hospital on October 5, 1963. The infant was born prematurely weighing 1700 grams. A transient hyperbilirubinemia occurred in the neonatal period. At two months of age a percutaneous liver biopsy was obtained because of icterus and hepatosplenomegaly and demonstrated active cirrhosis with bile stasis. Physical examination revealed slight icterus with hepatosplenomegaly and a prominent venous pattern over the anterior chest and upper abdomen. Laboratory studies revealed: hemoglobin 8.5 g/100 cc; white blood cells 6,100; total serum bilirubin 3 mg/dl; serum alkaline phosphatase 27 Bodansky units; SGOT 165 units; SGPT 85 units; prothrombin time 100%; urinalysis bile positive. A barium esophagram was normal.

Course

For suspected biliary atresia an exploratory laparotomy was recommended but consent for operation was refused by the infant's parents. A percutaneous liver biopsy demonstrated "postnecrotic cirrhosis." The infant had massive gastrointestinal bleeding at 14 months of age which stopped spontaneously. The infant died at 17 months of age with massive ascites and respiratory distress. Autopsy examination disclosed a cirrhotic liver and a choledochal cyst measuring 10 × 13 × 15 cm, compressing the portal vein and with changes consistent with portal hypertension.

Case 2. A 14-year-old girl was referred to Children's Hospital March 3, 1969, because of massive hematemesis and an enlarging abdomen of two years duration. A barium esophagram obtained elsewhere suggested esophageal varices. Physical examination revealed a healthy teenage girl with splenomegaly. Laboratory studies were: hemoglobin 11.5 g/100 cc; white blood cells 3,100; platelet count 61,000; prothrombin time 100%; total serum bilirubin 1.6 mg/dl; serum alkaline phosphatase 6.8 Bodansky units; SGOT 72 units; SGPT 58 units; total serum protein 6.3 g/100 cc; serum globulin 2.6 g/100 cc; serum albumin 3.7 g/cc.

Course

A percutaneous splenoportogram demonstrated esophageal varices with no filling of the portal vein. The splenic pulp pressure measured 490 mm water. At operation a choledochal cyst measuring 15 cm in diameter and containing 1.5 l of bile was encountered. It obstructed the portal vein by extrensic compression. A Roux-en-Y choledochocystojejunostomy was performed. An operative portoportogram following decompression of the choledochal cyst revealed a patent portal vein and portal pressure of 340 mm water. One month later the white blood count and platelet count had returned to normal. Four years following operation the patient was free of symptoms with normal hemogram and physical examination reported as normal by the family physician.

The cases previously recorded in the literature are summarized in Table 1.

Case 3. A 2-year-old girl with jaundice, acholic stools and hepatosplenomegaly had a splenectomy for bleeding esophageal varices. Because of recurrent gastrointestinal bleeding, abdominal exploration a month later revealed a large choledochal cyst and a choledochocystojejunostomy was performed. Three years later no varices could be found.[4]

Case 4. A 5-year-old girl with epistaxis, jaundice and hepatosplenomegaly had a liver biopsy which was diagnosed as a malignant cholangioma. Seven years later, an exploratory laparotomy for massive gastrointestinal bleeding revealed a large choledochal cyst and a choledochocystojejunostomy was done. Ten months later no varices were demonstrated.[4]

Case 5. A 5½-year-old girl with massive gastrointestinal bleeding from esophageal varices had a venous splenorenal shunt carried out. The expected fall in portal venous pressure did not occur following this procedure. On further exploration of the ab-

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Submitted for publication: March 8, 1979.

0003-4932/79/1100/0638 $00.60 © J. B. Lippincott Company

TABLE 1. *Choledochal Cyst with Portal Hypertension*

Authors	Age	Sex	Operation	Result	Follow-up
Gillis & Sargent	2 yr	Female	Choledochocystjejunostomy	Recovered	3 yr
Gillis & Sargent	5 yr	Female	Choledochocystjejunostomy	Recovered	10 mo
Fonkalsrud & Boles	5½ yr	Female	Splenorenal Shunt and Choledochocystjejunostomy	Recovered	1 yr
Duckett, Erachlis, Longino	6 yr	Female	None	Died	
Martin & Rowe	8 mo	Female	None—refused by parents	Died	
Martin & Rowe	2 yr	Female	Choledochocystjejunostomy	Recovered	3 yr
Martin & Rowe	14 yr	Female	Choledochocystjejunostomy	Recovered	4 yr

domen, a choledochal cyst was found and a choledochocystojejunostomy was performed with an immediate fall in portal pressure. One year later esophageal varices were not demonstrated.[3]

Case 6. A 6-year-old girl was admitted with ascites and massive upper gastrointestinal bleeding. An erroneous diagnosis of malignant tumor was made on the basis of a smear of ascitic fluid and radiotherapy was administered. The child later died and postmortem examination revealed a large choledochal cyst without evidence of malignancy.[2]

Discussion

A choledochal cyst is classically associated with the triad of jaundice, abdominal mass, and pain. As emphasized by previous authors,[5,6] however, pain is a late and inconsistent finding. Since four of the six children presented with gastrointestinal bleeding, this symptom should be added to the list of presenting complaints of choledochal cyst.

Two mechanisms have been proposed by which a choledochal cyst may lead to portal hypertension: by direct compression of the portal vein; or, by extrahepatic biliary obstruction with the subsequent development of biliary cirrhosis leading to portal hypertension.[4] It would appear from our experience that direct compression of the portal vein is the most common mechanism.

Choledochal cyst as a cause of portal hypertension, albeit rare, should be considered a possibility in all instances of bleeding esophageal varices. A careful roentgenographic contrast study of the duodenum will generally demonstrate displacement and compression of the duodenum. In the absence of clinical jaundice, the choledochal cyst itself may be demonstrated by contrast media administered orally or intravenously. In the event of jaundice, transhepatic cholangiography has been suggested but was not employed in any of our patients.

In three of the six patients, needle biopsy of the liver was performed. In each instance, the findings were sufficiently varied and unusual as to lead to a misleading diagnosis even when interpreted by nationally recognized authorities in pediatric pathology.

Our experience combined with that of others indicates that successful decompression of the choledochal cyst affords satisfactory relief of the portal hypertension in that it was successful in all four patients in which it was performed. The creation of a portal-systemic shunt has not been necessary. Measurement of portal pressure both before and after decompression of the choledochal cyst and a before and after portoportogram afforded reliable and documentary evidence of adequate portal decompression.

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